Eyes - giant cell arteritis

Summary

- Giant cell arteritis is inflammation of the arteries that can cause sudden blindness in one or both eyes.
- New onset headache and vision loss are the most common symptoms.
- People over the age of 50 years are at risk of developing the disease, for reasons unknown.
- Treatment includes immediate high-dose corticosteroids, which can provide symptom relief within 48 to 72 hours.

Giant cell arteritis is a particular kind of inflammation of the arteries that requires urgent treatment. The inflammation causes the artery to narrow, which reduces the blood supply to the area. In severe cases, the blood vessel closes completely. Although any medium or large-sized artery can develop this condition, those of the temples are most commonly targeted. Giant cell arteritis is also known as temporal arteritis.

If the blood vessels servicing the eyes are affected, sudden blindness in one or both eyes can result. This vision loss is usually severe and permanent. Around one person in 500 experiences giant cell arteritis, with twice as many women affected as men. The average age at diagnosis is 70 years. There is no cure, but prompt treatment with corticosteroids can relieve the symptoms and usually, but not always, prevent vision loss in the other eye if it is not yet involved.

Symptoms of giant cell arteritis

The symptoms of giant cell arteritis can include:

- Fever
- Generally feeling unwell
- Intermittent or constant headaches that can vary from mild to severe
- Jaw or tongue pain when chewing or talking
- Tender temples – for example, hair brushing may hurt
- Red, inflamed and painful patches on the scalp, including localised hair loss (alopecia)
- Death of affected areas of scalp (necrosis) – in severe cases of alopecia
- Sudden double vision (diplopia)
- Other vision problems such as flashing lights, colour changes and blurring
- Sudden and painless blindness in one eye
- Sudden and painless blindness in the other eye – this may occur one to 10 days later.

Causes of giant cell arteritis

‘Giant cells’ are large cells with many nuclei. The causes of giant cell arteritis are unknown. Some researchers believe that it may be a type of autoimmune disorder. Risk factors include:

- Gender – twice as many women as men are affected.
- Age – people over the age of 50 years are more susceptible. The average age at diagnosis is 70 years.
- Race – giant cell arteritis is more common among Caucasians.
- Region – prevalence is higher in northern European nations.

The link to polymyalgia rheumatica

About half of all people with giant cell arteritis are diagnosed with polymyalgia rheumatica. This is a collection of symptoms rather than a specific disease. Typical symptoms of polymyalgia rheumatica include:
Fatigue
Depression
Fever
Night sweats
Weight loss
Muscle aches
Stiffness in the neck, shoulders and hips.

It is thought that these symptoms are triggered by some kind of inflammatory response in the body. Since giant cell arteritis and polymyalgia rheumatica often occur at the same time, some researchers suspect they may be caused by the same underlying disease.

**Diagnosis of giant cell arteritis**

Giant cell arteritis is diagnosed using a number of tests including:

- **Medical history** – to check for the presence of risk factors. The doctor will strongly suspect giant cell arteritis if the person is aged 65 years or more.
- **Physical examination** – for example, the doctor may look for alopecia, scalp lesions, tenderness and a reduced pulse in the temporal arteries.
- **Eye examination** – if the eye is affected, the optic disc looks pale and puffy. This condition is known as anterior ischaemic optic neuropathy and is the worst complication of giant cell arteritis.
- **Blood test** – some blood tests flag the possible presence of giant cell arteritis or polymyalgia rheumatica. A raised erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) test may be an indicator.
- **Other tests** – these may be used to rule out other diseases that may present with similar symptoms – for example, diabetes, atherosclerosis, thyroid dysfunction, trigeminal neuralgia, dermatomyositis, multiple myeloma or rheumatoid arthritis.
- **Biopsy of the artery** – a small piece of artery is removed and examined microscopically in a laboratory. An artery affected by giant cell arteritis is inflamed, narrowed and shows a particular pattern of immune cells. Giant cells are not always found. Biopsy confirms about 94 per cent of cases so it is an important test to confirm the diagnosis. Both temporal arteries may need to be biopsied.

**Treatment of giant cell arteritis**

Treatment aims to stop any more damage to the affected tissues. If giant cell arteritis is suspected, doctors will start immediate drug treatment before the results of the biopsy come back or even before the biopsy can be arranged. Quick action is needed to preserve the person’s remaining sight.

Treatment options will include:

- **Corticosteroid drugs** – this is the preferred treatment because it can provide symptom relief within 48 to 72 hours and reduce the risk of future vision loss. Once the symptoms are managed, the dosage can be gradually reduced over a period of weeks to low-dose maintenance levels. In severe cases with vision loss, the person may undergo intravenous treatments for the first five or so days, and then continue with oral corticosteroids. Additional medications, such as methotrexate, may be added to help decrease the corticosteroid dose once the inflammation is controlled.
- **Drug treatment to prevent osteoporosis** – long-term use of corticosteroids weakens bones and known side effects include osteoporosis, fractures and bone infections. Other medications may be given to reduce bone loss.
- **Ongoing medical supervision** – drug therapy must be carefully monitored to maximise symptom relief and minimise bone loss. Steroids will usually cure this condition if the treatment is given for long enough – usually 12 to 18 months. Giant cell arteritis tends to resolve itself within five years. Relapses occur most often in the first 18 months of treatment or within one year of completing treatment. About one in four people experience a relapse but it is impossible to know in advance which of those affected are at risk.

**Where to get help**

- Your local eye hospital or emergency department